Giant lupus vulgaris: A rare presentation

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ABSTRACT

Cutaneous tuberculosis continues to be an important public health problem even with the availability of highly effective anti-tuberculous drugs. It constitutes 0.1% of all cases of extrapulmonary tuberculosis. Lupus vulgaris is the most common form of cutaneous tuberculosis that occurs in previously sensitized individuals with a moderate degree of immunity against tubercle bacilli. The different types of lupus vulgaris include plaque, ulcerative, vegetative, papular and nodular, and tumor forms. A 40-year-old man presented with large multiple plaques over right upper limb, right side of chest and back, and right lower limb for the past 30 years. Histopathology showed numerous noncaseating granulomas with Langhan's type of giant cells. The Mantoux test showed strong positivity and there was excellent response to anti-tuberculous treatment. This case is being reported because of its extreme chronicity of 30 years duration, unusually large size and multiplicity of lesions.

Key words: Cutaneous tuberculosis, giant plaque, lupus vulgaris

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INTRODUCTION

Cutaneous tuberculosis makes up a small proportion of extrapulmonary tuberculosis. Studies from India report an incidence of 0.1% of all cases of extrapulmonary tuberculosis.[1] It takes different clinical forms depending on patient's immune status.[2] Lupus vulgaris is the most common type of cutaneous tuberculosis, with most varied manifestations.[3] It is a chronic and progressive form occurring in individuals with high degree of tuberculin sensitivity and moderate immunity. A characteristic feature of lupus vulgaris is its extremely chronic course with slow but steady growth of the lesions over a period of many years, even decades. Head and neck are the sites commonly affected in European countries. In India, the buttocks, thighs, and legs are the common sites of involvement. Here we report a case of lupus vulgaris with very large sized, multiple plaques occurring at less common sites.

CASE REPORT

A 40-year-old man presented with large plaques of 30 years duration over right upper limb, right side of the chest and back, and right lower limb. The lesion started as a small papule over right upper limb near the elbow when he was 10 years old. Since then it steadily grew in size to involve whole of the limb and also the right side of the chest and back. He developed similar lesions, one over the right thigh after about 2 years of the first lesion that gradually enlarged to involve most of the thigh circumferentially and one more lesion over the left side of the back since one year [Figure 1]. The lesions were asymptomatic except for mild itching. There was no history of trauma prior to the onset of the lesions or the past history suggestive of tuberculosis of any part of the body. There was no family history of tuberculosis or contact with a tuberculosis patient. He consulted many doctors and applied various creams but of no help.

On examination, there were three plaques, two large and one small, with well-defined borders and irregular margins:

- Plaque no. 1: involving right upper limb, right side of the chest and back measuring about 60 × 45 cm.
- Plaque no. 2: about 6 × 3 cm over the left side of the back.
- Plaque no. 3: over right thigh, involving circumferentially, measuring about 40 cm longitudinally and 50 cm circumferentially.

Within the plaque, there were areas of thick hyperkeratosis and large, thick adherent crusts on an erythematous base. Ulceration at some places and areas of scarring and atrophy in

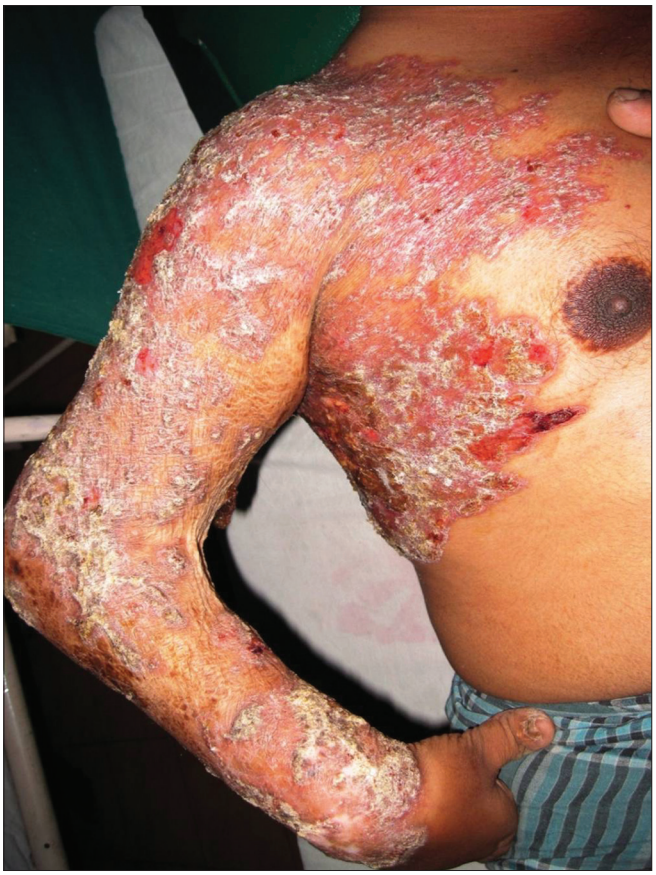

Figure 1: Clinical photograph showing a large plaque over right upper limb and right side of the chest before treatment

between the areas of hyperkeratosis were present. There was no regional or generalized lymphadenopathy. The patient could not extend the shoulder and elbow completely because of contracture. Systemic examination was normal.

Hematological investigations revealed anemia (Hb: 10.1 g%), raised erythrocyte sedimentation rate (67 mm/first hour). The Mantoux test was strongly positive with a reading of 22 × 20 mm at 72 h. Chest X-ray was normal. X-ray of the involved parts showed no evidence of underlying tissue infiltration. Sputum was negative for acid fast bacillus (AFB). Scrapings from crusts were negative for fungus. Culture for *M. tuberculosis* and fungus were negative. ELISA for HIV was nonreactive. Histopathologic examination of the biopsy specimen revealed hyperplastic epidermis, upper and mid-dermis showing numerous noncaseating epitheloid granulomas with Langhan's type of giant cells [Figure 2]. No fungal elements or AFB were seen on tissue section.

Based on clinical features and investigations, a diagnosis of lupus vulgaris was made and the patient was started on antitubercular treatment cat-I. There was dramatic improvement in skin lesions within 3 weeks and after 6 weeks the lesions showed 70–80% of improvement. After 12 weeks, there was almost total healing of the lesions [Figure 3].

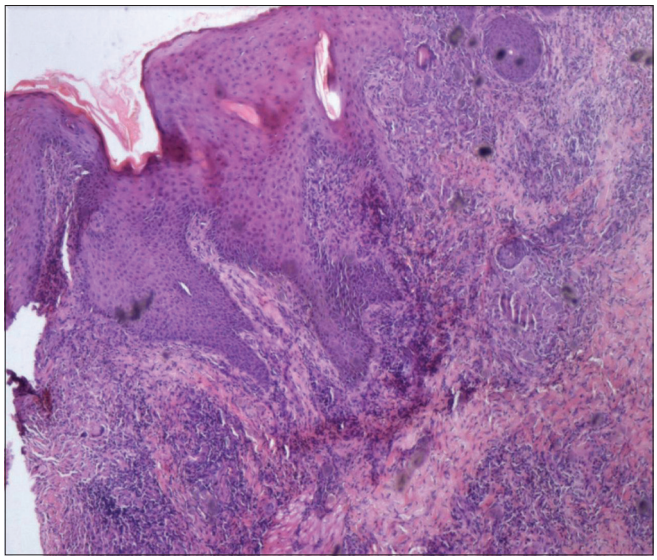

Figure 2: Skin biopsy showing hyperplastic epidermis, upper and mid dermis showing noncaseating epitheloid granulomas with Langhan's type of giant cells

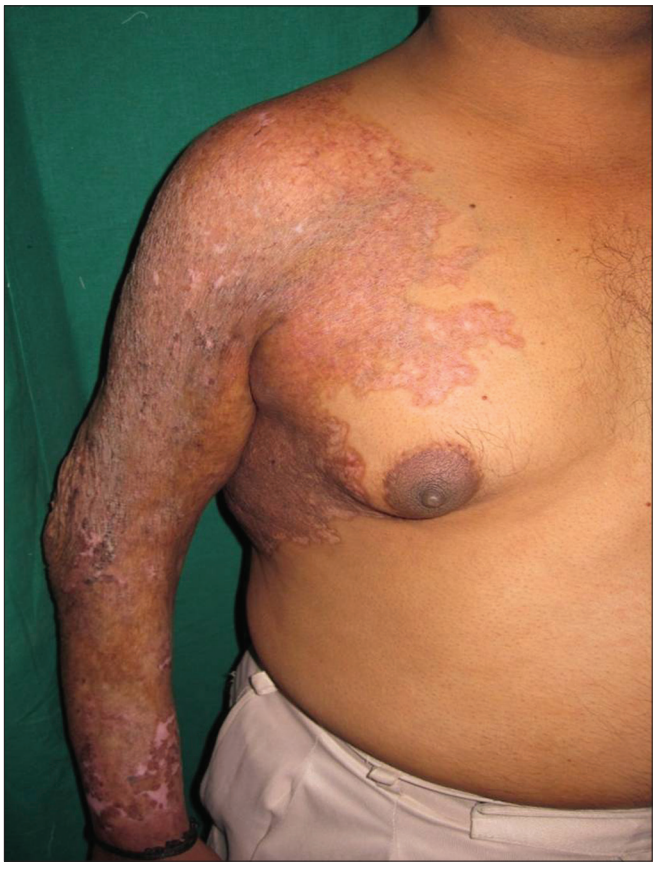

Figure 3: Clinical photograph after 12 weeks of ATT showing healing with scarring and atrophy

DISCUSSION

Lupus vulgaris is an extremely chronic, progressive form

of cutaneous tuberculosis. The earliest description of lupus vulgaris was given by Erasmus Wilson in 1865.[4] It usually occurs through contiguous extension of the disease from underlying affected tissue or hematogenous or lymphatic spread.[5] It can also arise after exogenous inoculation or as a complication of BCG vaccination.[6] Lupus vulgaris is the most common type of cutaneous tuberculosis in India and commonly seen in the lower half of the body involving legs, thighs, buttocks, and feet.[7] It is attributed to the habit of children playing without clothing or shoes and defecating in the open.[8] Pyogenic infection of the gluteal region is also common in India and the breach in the integrity of the skin can serve as a portal of entry for the AFB.

The initial lesion is a small, reddish-brown papule or nodule which later forms larger plaques by peripheral enlargement and coalescence that show evidence of healing and scarring in one area and activity in another.[8] Atrophic scarring, with or without ulceration, is a prominent feature of lupus vulgaris.[2] The common variants are: plaque form, ulcerative and mutilating form, vegetative form, papular and nodular form and tumor-like (hypertrophic) form. The plaque form is the common type that presents as flat plaques with irregular or serpigenous edges, surface may be smooth or covered with psoriasiform scale. Large plaques may show irregular areas of scarring with islands of active lupus tissue, the edge may be thickened and hyperkeratotic.[5] Histopathology shows tuberculoid granulomas composed of lymphocytes, plasma cells, epitheloid cells and giant cells, scant or absent central caseation, in the superficial dermis. The epidermis is usually hyperplastic, but may be atrophic or ulcerated. Tubercle bacilli are difficult to demonstrate.[3]

After a thorough search of the literature, the largest size of lupus vulgaris lesion we could find was 30 × 25 cm.[2] Ours may be the first case showing largest lesions measuring 60 × 45 cm and 40 × 50 cm. Other special features in our case were multiplicity (LV commonly presents as a single lesion) and extreme chronicity of the lesions, and occurrence at less common sites such as upper limb, chest, and back. This case also shows the level of ignorance among patients and consequent failure to take proper anti-tuberculous treatment despite extensive campaign in print and audiovisual media.

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